

What Is Claimed:

1 1. A method for treating a subject suffering
2 from phenylketonuria and/or phenylalanemia, said method
3 comprising:

4 enterally administering to the subject a LNAA
5 supplement in which the weight ratio of Leu to Val is
6 greater than 2:1.

1 2. A method according to claim 1, wherein the
2 LNAA supplement is substantially free from phenylalanine.

1 3. A method for treating a subject suffering
2 from phenylketonuria and/or phenylalanemia, said method
3 comprising:

4 enterally administering to the subject a LNAA
5 supplement in which the weight ratio of Leu to iLeu is
6 greater than 3:1.

1 4. A method according to claim 3, wherein the
2 weight ratio of Leu to Val in the LNAA supplement is
3 greater than 2:1.

1 5. A method according to claim 4, wherein the
2 LNAA supplement is substantially free from phenylalanine.

1 6. A method for treating a subject suffering
2 from phenylketonuria and/or phenylalanemia, said method
3 comprising:

4 enterally administering to the subject a LNAA
5 supplement which comprises one or more LNAAs and which
6 further comprises Lys.

1 7. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu.

1 8. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu and wherein the weight
3 ratio of Leu to iLeu in the LNAA supplement is greater
4 than about 0.5:1.

1 9. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu and wherein the weight
3 ratio of Leu to iLeu in the LNAA supplement is greater
4 than 3:1.

1 10. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu and wherein the weight
3 ratio of Leu to Val in the LNAA supplement is greater
4 than about 0.5:1.

1 11. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu and wherein the weight
3 ratio of Leu to Val in the LNAA supplement is greater
4 than 2:1.

1 12. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu; wherein the weight ratio
3 of Leu to iLeu in the LNAA supplement is greater
4 than about 0.5:1; and wherein the weight ratio of Leu to
5 Val in the LNAA supplement is greater than about 0.5:1.

1 13. A method according to claim 12, wherein
2 the weight ratio of Leu to iLeu in the LNAA supplement is
3 greater than 3:1.

1 14. A method according to claim 12, wherein
2 the weight ratio of Leu to Val in the LNAA supplement is
3 greater than 2:1.

1 15. A method according to claim 6, wherein the
2 LNAA supplement comprises Leu; wherein the weight ratio
3 of Leu to iLeu in the LNAA supplement is greater than
4 3:1; and wherein the weight ratio of Leu to Val in the
5 LNAA supplement is greater than 2:1.

1 16. A method according to claim 6, wherein the
2 LNAA supplement is substantially free from phenylalanine.

1 17. A LNAA supplement comprising Leu and Val
2 in which the weight ratio of Leu to Val is greater than
3 2:1.

1 18. A LNAA supplement according to claim 17,
2 wherein the LNAA supplement is substantially free from
3 phenylalanine.

1 19. A LNAA supplement comprising Leu and iLeu
2 in which the weight ratio of Leu to iLeu is greater than
3 3:1.

1 20. A LNAA supplement according to claim 19,
2 wherein the weight ratio of Leu to Val in the LNAA
3 supplement is greater than 2:1.

1 21. A LNAA supplement according to claim 20,
2 wherein the LNAA supplement is substantially free from
3 phenylalanine.

1 22. A LNAA supplement comprising one or more
2 LNAAs and further comprising Lys.

1 23. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu.

1 24. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu and wherein the
3 weight ratio of Leu to iLeu in the LNAA supplement is
4 greater than about 0.5:1.

1 25. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu and wherein the
3 weight ratio of Leu to iLeu in the LNAA supplement is
4 greater than 3:1.

1 26. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu and wherein the
3 weight ratio of Leu to Val in the LNAA supplement is
4 greater than about 0.5:1.

1 27. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu and wherein the
3 weight ratio of Leu to Val in the LNAA supplement is
4 greater than 2:1.

1 28. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu; wherein the
3 weight ratio of Leu to iLeu in the LNAA supplement is
4 greater than about 0.5:1; and wherein the weight ratio of
5 Leu to Val in the LNAA supplement is greater than about
6 0.5:1.

1 29. A LNAA supplement according to claim 28,
2 wherein the weight ratio of Leu to iLeu in the LNAA
3 supplement is greater than 3:1.

1 30. A LNAA supplement according to claim 28,
2 wherein the weight ratio of Leu to Val in the LNAA
3 supplement is greater than 2:1.

1 31. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises Leu; wherein the
3 weight ratio of Leu to iLeu in the LNAA supplement is
4 greater than 3:1; and wherein the weight ratio of Leu to
5 Val in the LNAA supplement is greater than 2:1.

1 32. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement is substantially free from
3 phenylalanine.

1 33. A LNAA supplement according to claim 22,
2 wherein the LNAA supplement comprises, per 500 mg of LNAA
3 supplement:
4 from about 100 mg to about 290 mg of Tyr;
5 from about 25 mg to about 75 mg of Trp;
6 from about 15 mg to about 50 mg of Met;
7 from about 15 mg to about 55 mg of iLeu;
8 from about 15 mg to about 50 mg of Threo;
9 from about 15 mg to about 55 mg of Val;
10 from about 15 mg to about 200 mg of Leu;
11 from about 10 mg to about 30 mg of His; and
12 from about 5 mg to about 200 mg of Lys.

1 34. A LNAA supplement according to claim 33,
2 wherein the LNAA supplement comprises, per 500 mg of LNAA
3 supplement, from about 10 mg to about 30 mg of Lys.

1 35. A LNAA supplement according to claim 33,
2 wherein the LNAA supplement is substantially free from
3 arginine.

1 36. A LNAA supplement according to claim 33,
2 wherein the LNAA supplement is substantially free from
3 phenylalanine.

1 37. A LNAA supplement comprising, per 600 mg
2 of LNAA supplement:
3 from about 100 mg to about 290 mg of Tyr;
4 from about 30 mg to about 90 mg of Trp;
5 from about 25 mg to about 75 mg of Met;
6 from about 15 mg to about 45 mg of iLeu;
7 from about 15 mg to about 50 mg of Threo;
8 from about 15 mg to about 50 mg of Val;
9 from about 40 mg to about 200 mg of Leu;
10 from about 15 mg to about 45 mg of His; and
11 from about 15 mg to about 50 mg of Arg.

1 38. A LNAA supplement according to claim 37,
2 wherein the LNAA supplement further comprises Lys.

1 39. A LNAA supplement according to claim 37,
2 wherein the LNAA supplement further comprises, per 600 mg
3 of LNAA supplement, from about 5 mg to about 200 mg of
4 Lys.

1 40. A LNAA supplement according to claim 37,
2 wherein the LNAA supplement is substantially free from
3 phenylalanine.

1 41. A method for treating a subject suffering
2 from a condition involving a metabolic disorder involving
3 the metabolism of a first amino acid X, said method
4 comprising:
5 enterally administering to the subject a
6 composition which is substantially free from said first
7 amino acid X and which comprises a second amino acid Y
8 that competes with amino acid X at a gastrointestinal
9 tract transporter.

1 42. A method according to claim 41, wherein
2 the condition is not phenylketonuria and/or
3 phenylalanemia.

1 43. A method according to claim 42, wherein
2 the gastrointestinal tract transporter is a Caco-2 cell
3 transporter.

1 44. A method according to claim 41, wherein
2 the gastrointestinal tract transporter is a Caco-2 cell
3 transporter.

1 45. A method according to claim 44, wherein
2 the condition is tyrosinemia; wherein the first amino
3 acid X is tyrosine; and wherein the second amino acid Y
4 is selected from Phe, Leu, Trp, Lys, His, and
5 combinations thereof.

1 46. A method according to claim 44, wherein
2 the condition is tyrosinemia; wherein the first amino
3 acid X is selected from phenylalanine, tyrosine, and
4 combinations thereof; and wherein the second amino acid Y
5 is selected from Leu, Trp, Lys, His, and combinations
6 thereof.

1 47. A method according to claim 44, wherein
2 the condition is alkaptonuria; wherein the first amino
3 acid X is selected from phenylalanine, tyrosine, and
4 combinations thereof; and wherein the second amino acid Y
5 is selected from Leu, Trp, Lys, His, and combinations
6 thereof.

1 48. A method according to claim 44, wherein
2 the condition is homocystinuria; wherein the first amino
3 acid X is methionine; and wherein the second amino acid Y
4 is an amino acid that competes with methionine at a
5 gastrointestinal tract transporter.

1 49. A method according to claim 44, wherein
2 the condition is a disorder affecting metabolism of a
3 branched amino acid selected from leucine, isoleucine,
4 valine, and combinations thereof; wherein the first amino
5 acid X is selected from leucine, isoleucine, valine, and
6 combinations thereof; and wherein the second amino acid Y
7 is an amino acid that competes with the first amino acid
8 X at a gastrointestinal tract transporter.

1 50. A method according to claim 49, wherein
2 the condition is selected from maple syrup urine disease,
3 isovaleric acidemia, methylmalonic acidemia, and
4 propionic acidemia.

1 51. A method according to claim 41, wherein
2 said method further comprises:
3 restricting the subject's dietary intake of the
4 first amino acid X.

1 52. A method according to claim 41, wherein
2 said method further comprises:
3 not restricting the subject's dietary intake of
4 the first amino acid X.

1 53. A method according to claim 52, wherein
2 said enteral administration is carried out substantially
3 at mealtime.

1 54. A method according to claim 41, wherein
2 said enteral administration is carried out substantially
3 at mealtime.

1 55. A method according to claim 41, wherein
2 said enteral administration is carried out orally.